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THE ASSOCIATION OF PURPURA AND ACUTE CIRCUMSCRIBED  
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H ENOCH<sup>2</sup> and, later, Couty<sup>3</sup> were the first writers to call attention to a form of purpura characterized by recurring attacks of cutaneous hemorrhage, associated with gastro-intestinal disturbance and acute œdema of the skin and subcutaneous tissue. There were frequently present, in addition, pains in the joints together with articular effusion and extra-articular œdema. Couty, by collecting and analyzing a considerable number of cases, came to the conclusion that this class of cases should be separated from all varieties of purpura. This form is characterized by a brusque onset, and the several symptoms are inclined to come in distinct attacks. Of these cases there is a great divergence in the prominence of the several symptoms. In some, the cutaneous œdema is the chief feature, the purpuric eruption holding a secondary place. Sudden attacks of acute swelling of a limb or part of a limb occur, which quickly subside to be followed by other attacks of the same nature. In other cases the cutaneous disturbance is almost nul, the only feature of prominence being successive crops of purpuric patches, with vomiting and diarrhœas occurring often in the intervals between the outbreaks of purpura. These gastro-intestinal disturbances may in their turn constitute the salient fea-

<sup>1</sup> Read before the American Dermatological Association, at New London, September, 1892.

<sup>2</sup> Berlin, Klin, Wochenschrift, 1874.

<sup>3</sup> Gazette Hebdomadaire, 1876.

ture, the patient suffering from severe attacks of colic with nausea and vomiting, while the pathological appearances on the cutaneous surfaces are very slight. Couty considers, therefore, that there are four sub-divisions of this form of purpura: one, the purpura is complicated with cutaneous œdema and gastro-intestinal crises; two, the purpura is complicated with œdema; three, the purpura is only accompanied by intestinal troubles; and four, the purpura exists alone.

Some later writers are unable to see the necessity of separating these forms from the classical purpura. Immermann, in *Ziemssen's Encyclopedia*, remarks "we cannot understand why Hænoch should be inclined regard to such cases as constituting a special form of disease entirely distinct from morbus maculosus," and Atkinson in *Pepper's System* is of the same mind. Latterly, Osler<sup>1</sup> has published two cases in an article entitled "A Form of Purpura." The characteristics of this form are first, recurring outbreaks of purpura often associated with urticaria or local œdema: second, articular pain sometimes with swelling; third, gastro-intestinal disturbance with vomiting, diarrhœa and occasionally hemorrhage; fourth, hematuria, albuminuria, and sometimes a fatal nephritis. The first of his cases occurred in a boy of six who was seized with attacks of colic and diarrhœa accompanied by an urticarial eruption tending to become purpuric, and by moderate hemorrhage from the bowels. Later, œdema of the ankles and legs appeared and the boy died in six weeks of Bright's disease. The second case, a man of forty-six, began with pain in the arms and knees, with œdema and purpuric patches of the skin together with severe paroxysms of colic and diarrhœa, but no blood in the stools. Albumen and blood were passed in the urine. He recovered in a few days. Osler considers that these cases belong under purpura rheumatica of which they constitute the most aggravated and serious form. The modern French writers as Brocq and Besnier in dealing with this subject refer to an article on purpura in the *Dictionnaire des Sciences Médicales*, by Albert Mathieu, as representing the best exposition of the subject. Mathieu includes the form under consideration in his purpura exanthématique rhumatoïde a type that ranges from purpura simplex, limited to the lower limbs and accompanied perhaps with slight articular pains, to the cases of general purpuric eruption with internal hemorrhage and severe articular symptoms.

<sup>1</sup> N. Y. Med. Journal. Dec. 22, 1888.



Gastro-intestinal disturbances and cutaneous manifestations in the form of multiform erythema, urticaria, erythema nodosum and acute œdema, are regarded as symptoms belonging to this type, although very variable in their occurrence; and thus the form of Couty is also included under this general heading. The acute fulminating forms with high fever and typhoidal symptoms, Mathieu classifies as purpuras infectieux.

Two cases that were accompanied by purpuric manifestations have lately come under my observation which I will briefly describe.

Case 1. A boy one and a half years old, well nourished. No family history of importance could be elicited. Had been well until two days previously, when the mother noticed in the evening that the left ear was considerably swollen and also the left lower leg and the right fore-arm. Over the affected areas a few purpuric patches could be seen. The child was fretful and uncomfortable. Two days later, when seen at the Carney Hospital, the œdema of the ear had disappeared. The left lower leg was the seat of a very marked œdema, without redness or inflammatory appearances. The œdema extended from the knee to the ankle, where it ended quite abruptly. The skin of the affected part was quite tense and the œdema firm and hard, pitting only slightly on pressure. Numerous small hemorrhagic spots were seen in the œdematous area. Upon the right fore-arm was a similar œdema, very tense and rising to a considerable height at the centre, covered also with small purpuric patches. The left fore-arm was normal, but at the base of the left thumb there was another sharply elevated area of acute œdema. The child was restless, bowels regular but stools said to be black and offensive. The next day the œdema had almost entirely disappeared from the left leg and right arm, while the right leg and foot, normal the previous day, were the seat of an enormous œdema covered with purpuric spots. The left fore-arm also had become greatly swollen over part of its extent. A few hemorrhagic spots were noticed on other parts of the trunk and extremities and those seen the previous day were much less prominent. There was some swelling and redness of the hard palate. The child had a good appetite, taking plenty of milk. The bowels moved two or three times, the stools being reported to be "coal black"; otherwise there were no hemorrhages from mucous surfaces. The temperature had oscillated between 100 and 101 degrees F.; the pulse was about 120. The child was not seen again as it was taken away from the hospital against advice.



Case 2. Was seen at Lynn with Dr. H. W. Newhall. The patient, a stout well nourished girl of seventeen, twelve days previously was seized with a sudden, intense pain in the calf of the leg, which continued for several days. Next came an attack of vomiting and gastric pain, and since has had similar attacks at intervals, besides severe pain in various joints without appreciable swelling. There had been, however, a very pronounced acute swelling of the whole back of the left hand which had subsided rapidly. Six days ago an eruption was first noticed on the buttocks, and later on the legs and body. When seen the face was pretty well covered with an erythematous eruption, bright red and slightly raised, suggesting measles somewhat. On the ears were one or two hemorrhagic patches with bullous lesions on the surface. On the upper part of the chest and back was a fainter eruption of an erythematous and in places urticarial character. Very little eruption on the abdomen. The buttocks were the seat of very marked cutaneous disturbance, being covered with hemorrhagic spots, upon some of which bullæ had formed, which were filled with blood. Others had been broken by scratching and blood crusts had formed. On the thighs there was very little cutaneous change, but on the legs and soles of the feet there were numerous purpuric patches. The extensor surfaces of the hand, especially the knuckles, were covered with purpuric patches and hemorrhagic bullæ. There was a moderate fever, about 101 degrees F. There were no hemorrhages from the mucous surfaces. The patient made a good recovery, the disease having apparently reached its acme when seen.

It is to be noted that in the first of these cases the chief symptom was the acute circumscribed œdema, the purpura being quite insignificant by comparison. In this case there were no gastro-intestinal disturbances, with the exception of probably slight hemorrhages from the bowels. In Case 2, the striking feature was the purpuric and bullous eruption, almost multi-form in character, as it appeared combined with urticarial and erythematous patches. There was marked colic coming on in paroxysms. There was one attack of acute circumscribed œdema.

There can be no doubt that these two cases are instances of the type of disease described by Henoch and Couty as purpura of nervous origin.

Turning now to acute angioneurotic, or circumscribed œdema. First described by Milton under the name of giant urticaria, it

has received attention of late at the hands of Quinke,<sup>1</sup> Riehl,<sup>2</sup> Strübing,<sup>3</sup> and quite recently of Joseph<sup>4</sup> and of Osler.<sup>5</sup> Its characteristics are well known to all dermatologists, and I think all will agree that it is not so uncommon as many of the writers of late have assumed. Its etiology remains obscure, although the general opinion prevails that it is produced through the agency of the nerves, and for this reason it has been called angioneurotic. It has been abundantly shown that in some cases the affection is hereditary. This was asserted first by Quinke, and Osler has published a remarkable instance of its persistence in the members of one family through five generations. In these cases there were also attacks of nausea, vomiting and diarrhœa. The association of its occurrence with the menstrual and climacteric periods has also been noted. Banke,<sup>6</sup> the director of a water cure establishment, has recently published an article in which he reports two cases of acute œdema in patients affected with cerebral and spinal irritation. In one the occurrence was often coincident with the menses. Traumatism may cause sudden and alarming attacks of œdema in the neighborhood of the part violated, as in three cases published by Horwitz.<sup>7</sup> Over indulgence in alcohol has been proved in a case of Joseph's to furnish the exciting cause of these attacks. In other cases there have been articular pains and swellings at the time of the outbreaks. Attacks of colic, nausea, and vomiting have frequently accompanied the œdematous swellings, a fact that has often been emphasized.

The association of acute cutaneous œdema with hemorrhages in various places is the point to which I wish to call especial attention. Joseph<sup>8</sup> has related before the congress of Prague a case of acute circumscribed œdema of the skin with paroxysmal hæmoglobinuria. Dr. J. G. Mumford<sup>9</sup> reports a case of a man who had been a bleeder and had had attacks of purpura and dangerous epistaxis, who developed an enormous swelling of the jaw upon the right side, which threatened suffocation, and finally subsided after opening into the mouth. Large clots and

<sup>1</sup> Monatsheft. f. prakt. Dermatologie, 1882.

<sup>2</sup> Wiener Med. Presse, 1888.

<sup>3</sup> Zeitschrift f. Klin. Med., 1885, IX.

<sup>4</sup> Ergänzungsheft zum Archiv, f. Dem. u. Syph., 1889.

<sup>5</sup> Am. Journal of Med. Sciences, 1888, Vol. II.

<sup>6</sup> Berlin. Klin. Wochenschr. 1892, No. 6.

<sup>7</sup> Phil. Med. News, Apr. 16, 1892.

<sup>8</sup> l. c.

<sup>9</sup> Boston Med. and Surg. Journal, March 5, 1891.



a great deal of blood were discharged from the opening. A further instance of the association of cutaneous hemorrhage and acute œdema has lately come under my own observation. A girl fourteen years of age was seen with purpuric patches over the lower legs with a few urticarial lesions interspersed. There was considerable pain in the joints. According to the mother's story many of the wheals on their disappearance leave purpuric spots. Two years previously the child had a similar attack lasting five months, accompanied by sudden acute swellings reaching an enormous size over various parts of the body, which always subsided quickly. A case is reported in the *Boston Medical and Surgical Journal*, by Dr. W. H. Holmes<sup>1</sup> entitled "A Case of Acute Local Œdema Complicated with Purpura and Salivation." This case corresponds in most respects with the first case I have reported, and it is therefore to be classed with the cases of Hænoch and Couty. Here, too, as in my own case, the local œdema was the feature of greatest prominence and the hemorrhagic lesions slight. It is not strange that the affection should be described as an acute œdema, and it may well be demanded, why is it not to be regarded as an acute œdema and the purpura as a complication? It should be constantly borne in mind, I think, that both purpura and acute œdema are properly speaking symptoms merely, although it may be impossible in the present state of our knowledge regarding their pathology to avoid using these terms oftentimes as if they represented well defined affections.

Thus it seems possible that some of the cases of acute œdema, complicated with gastro-intestinal troubles, belong in the same class, or may have the same etiology as the nervous purpura of Hænoch and Couty. All they lack to be included in this class is the presence of another symptom, to however slight an extent, namely, hemorrhage into the skin. It has been abundantly shown, that the other symptoms that accompany this form of purpura, as gastro-intestinal disturbances and pain and swelling of the joints, occur in many of the cases described as angio-neurotic œdema. Urticaria and erythema are occasionally observed in connection with the form of purpura under consideration, as was well illustrated by the second of my cases; and in acute œdema there is sometimes a typical urticaria seen. The relationship of purpura and acute œdema has already been indicated, I find, by Osler, in his article on a form of purpura,

<sup>1</sup> May 14, 1891.



which has been referred to above. His cases, which represent the type of Henoch and Couty, he considers to belong under *purpura rheumatica*. He observes "Are these cases truly rheumatic, or is not the articular affection on which so much stress is laid analogous to what we see in hemophilia and scurvy?" It is difficult to escape from the former view in the presence of characteristic cases of *peliosis rheumatica* with endocarditis and pericarditis, and yet the close relationship and even interchangeability of certain of these cases of *purpura* with *urticaria*, with *erythema nodosum*, and with the *angioneurotic œdema*, favor the suggestion that the entire group may depend upon some poison,—an alkaloid, possibly, the result of faulty chylopoietic metabolism, which in varying doses in different constitutions, excites in one *urticaria*, in a second *peliosis rheumatica*, and in a third a fatal form of *purpura*. "

It seems useless at the present time to attempt a strict classification of the different forms of *purpura*. The various forms described as *purpura simplex*, *purpura hemorrhagica*, etc., merge into one another, and are complicated with other symptoms in the most puzzling way. The investigation of Letzerich and more especially of Kolb would make it probable that some of the more acute and fatal cases are due to the action of a bacillus and its products. Kolb's work in Berlin upon three fatal cases of the type described by Henoch as *purpura fulminans*, revealed the presence of a bacillus which produced when inoculated in pure culture upon a large number of animals, purpuric spots and general internal hemorrhages. It is interesting to note that the products of the parasites were also capable of producing these appearances, as cultures that had been sterilized and filtered gave the same results upon inoculation.

Cutaneous œdema also, it seems to me, should be looked upon as essentially a symptom and not always due to the same cause. Its relationship to *urticaria* is very close, and it is with little doubt produced through the media of the nervous system. Just what the mechanism is, remains purely speculative in the absence of more thorough knowledge of the action of the nerves on the cutaneous vessels.







